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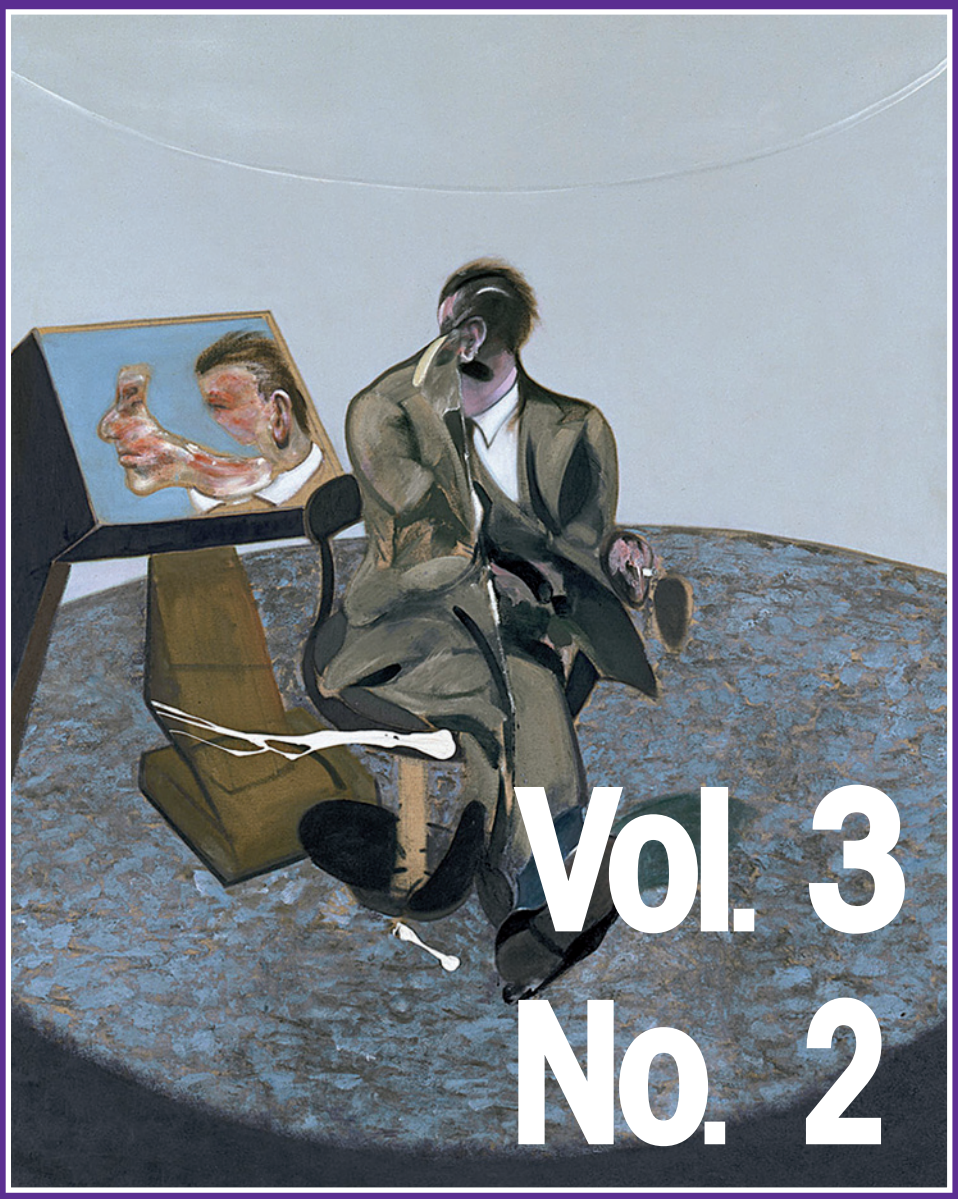

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Intratumoral Haemorrhage Causing an Unusual Clinical Presentation of a Vestibular Schwannoma

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SUMMARY – *We present a case of an elderly woman with no history of audiological disease with sudden onset of visual and hearing deficits associated with systemic clinical signs. On examination she had impairment of right CNs from V to X. CT and MR imaging demonstrated a cystic vestibular schwannoma with a rare intralesional fluid-fluid level correlated to a recent bleed. We include high quality MR images to show the acute impairment of the cranial nerves next to the tumour after acute bleeding. Our case report includes a voxel-based morphometry (VMB) analysis of the tumour that, as far as we know, has never been done before for such a tumour. VBM analysis was performed to calculate the hypothesized volume changes after the acute bleed which likely resulted in a sudden increase in the overall size of the tumour resulting in atypical clinical signs and symptoms due to the establishment of a mechanical conflict with the adjacent cranial nerves.*

Introduction

Vestibular schwannomas (VSs) are the most common (85%) cerebellar pontine angle (CPA) and intra acoustic channel (IAC) masses and account for about 8% of all brain tumours in adults: 90% are solitary and sporadic, the others are neurofibromatosis type 2 (NF2)-associated or multiple but unassociated with NF2 [1-4]. These benign encapsulated nerve sheath tumours are usually slow-growing solid masses but cystic changes as well as signs of previous bleeding have been reported at both MR imaging and histopathologic examination [5]; micro-haemorrhages have been implicated in the development of the cysts [6]. Cystic changes have been investigated as possibly responsible for bleeding (both intratumoral or subarachnoid) and there is general consensus on the importance of reporting these components [7]. Rarely an acute massive bleed can occur and this event can induce a sudden systemic illness and even death [8-13].

Case Report

We present a case of a 77-year-old hypertensive woman with acute onset of dizziness, headache, nausea, vomiting, sweating and diplopia occurring during defecation presenting slightly agitated with a rightward lateral gaze of the right eye.

After the first examination in the ER the patient underwent CT study (GE Medical System BrightSpeed 16) of the brain and a lesion causing mass effect on medulla oblongata, cerebellum and fourth ventricle without signs of hydrocephalus was demonstrated in the right CPA. The mass had mixed density because of a large hypodense cystic component and a hyperdense component due to an intralesional acute bleed with a small fluid/fluid level (hypodense/mildly hyperdense) in its declivous portion correlated to initial blood sedimentation. The diagnostic hypothesis of a giant vestibular schwannoma with "in tumour" haem-

orrhage was formulated because of a slight enlargement of the IAC and porus acusticus internus.

ENT examination disclosed a lack of contraction of the right masticatory muscles and ipsilateral face sensory dysfunction with almost absent corneal reflex on the right eye; a left beating nystagmus with horizontal and torsional components in primary position; Romberg's test positive on the right side. Audiometry showed right sensorineural hearing loss (SNHL) with a pure-tone average (PTA) of 75 dB and worse speech discrimination, that suggested for a retro-cochlear localization; Weber's test lateralized to the left side. Patient complained dysphagia and absence of pharyngeal reflex was observed at throat examination; flexible fiberoptic laryngoscopy was unremarkable.

The next day the patient underwent MR examination of the brain and CPA (GE Medical Systems Signa HDxt 1.5T) which confirmed the presence of a heterogeneous mass characterized by a solid component occupying the IAC (showing intermediate signal on all sequences and avidly enhancing on CE-T1w; Figure 1D) and a large cystic component (showing high signal on T2w and T2w-FLAIR; Figure 1A) extending into the PCA cistern. The hyperintensity of the cyst on T2w-FLAIR excluded an associated arachnoid cyst and the finding was correlated with a high level of protein (xanthochromic fluid). The fluid-fluid level described on CT was increased in size and was iso-hyperintense on T1w (Figure 1B) and hypointense on T2w-FLAIR (Figure 1A). This finding was correlated to the presence of mixed deoxyhaemoglobin and intracellular metahaemoglobin (acute/early-subacute haemorrhage; 12-72 hours/first several days). Into the cranial part of the tumour, close to the solid component, a small but strongly hypointense area was identified on T2*-GRE sequences (Figure 1C) and this finding was correlated with the presence of hemosiderin, indicating previous bleeding (>30 days). A fast imaging employing steady-state acquisition (FIESTA, GE Medical Systems high resolution steady-state free-precession sequence) clearly demonstrated the impairment of the right CNs V, VI, IX and X (Figure 2A-F). We hypothesized that the acute clinical presentation was caused by a sudden increase in the size of the cystic component of the tumour due to the acute bleeding which led to an acute impairment of adjacent cranial nerves. Although rare, this type of bleeding has been documented before [8-13] but a VBM analysis has never been reported in the literature. The VBM analysis was performed with ITK-SNAP 2.0 [14] (running on an independent workstation, Apple Mac Pro 1.1 – 2006) using the volumetric FIESTA sequence as reference. For more accurate results we chose a semiautomatic segmentation: first an automatic segmentation based on intensity regions was performed and a manual segmentation to improve the results of the automatic segmentation followed. Four different mass components were analyzed. 1) Solid: that part of the mass that showed intense enhancement on CE-T1w (those parts of the cystic wall less than 1 mm thick were not included); since only a few microcysts were found (cysts less than <1 mm in size), they were included in the solid component segmentation. 2) Cystic: including two millimetric cysts found inside the solid component of the tumour (about 3×1 mm and 5×2 mm on the axial plane) and the large cyst occupying the CPA cistern. 3) Fresh bleeding: the fluid declivous level related to recent bleeding was considered. 4) Old bleeding: the haemosiderin deposits found in the cranial part of the tumour. The result showed that the total tumour volume was more than 15 cm^3 (for accuracy 15616.63 mm^3) of which 8% solid (1257.41 mm^3), 65.68% cystic (10257 mm^3), 25.2% fresh bleeding (3943,78 mm^3) and only 1.07% hemosiderin (167.44 mm^3). Although an MR examination before the acute event was not available for comparison, these results are in favour of our hypothesis that a macro haemorrhage may be responsible for a significant sudden increase in the overall size of the mass (about 25% in our case) with the resulting clinical signs and symptoms due to the establishment of a mechanical conflict with the adjacent cranial nerves. This applies even more if we think how narrow the CPA cistern is.

Discussion

The typical clinical presentation of VS is a progressive uncomplicated unilateral SNHL possibly combined with chronic tinnitus and/or dizziness in otherwise healthy patients. Our results confirm that a macro-haemorrhage may be responsible for a significant sudden increase in the overall size of the mass with the resulting clinical signs and symptoms due to the establishment of an acute mechanical conflict with the adjacent cranial nerves. Unfortunately our patient refused surgery so we have no informa-

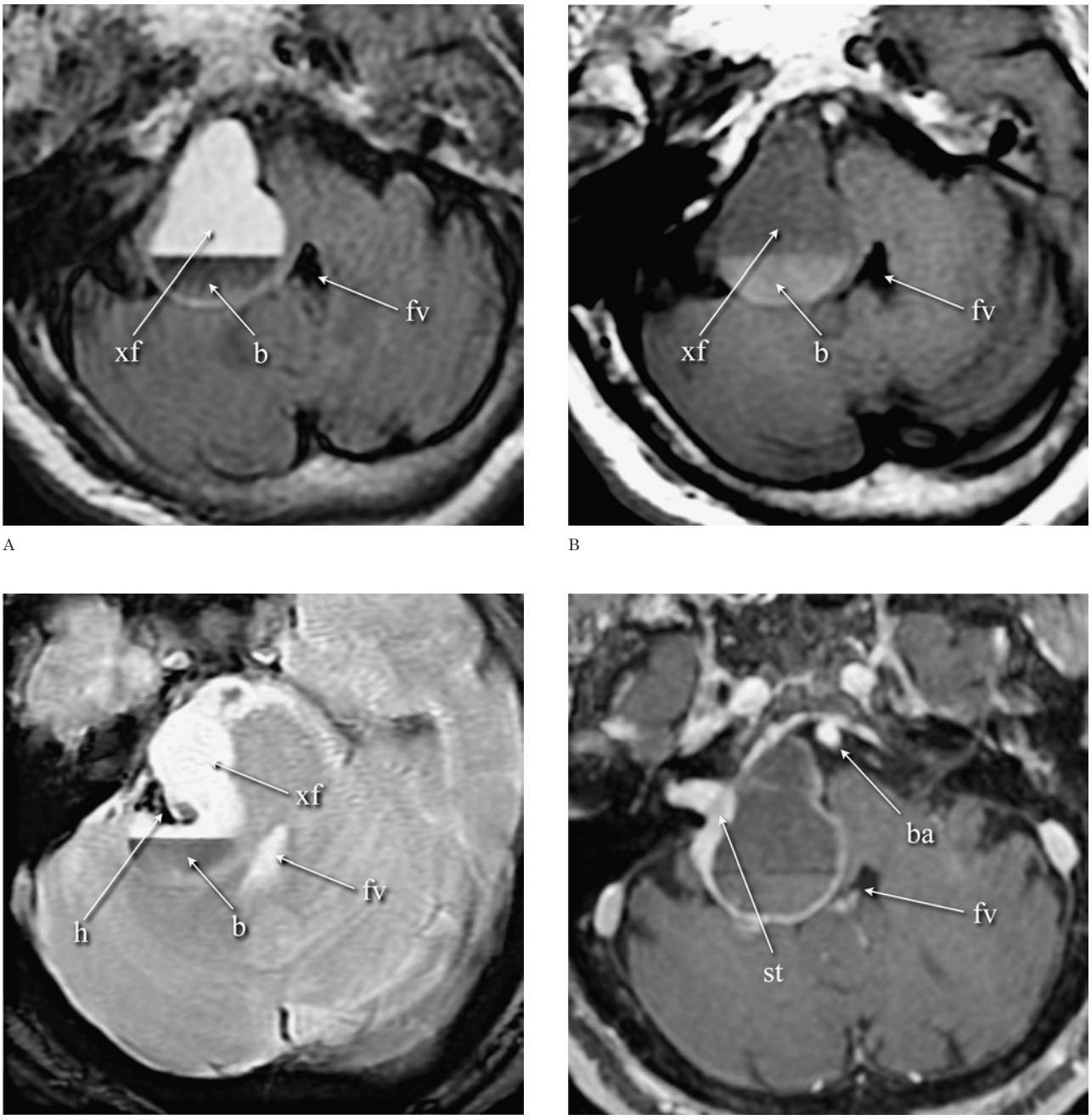

Figure 1 Axial T2w-FLAIR (A), T1w-TSE (B) and T2*-GRE (C). Axial multiplanar reconstruction (MPR) from a 3D T1w-FATSAT with Gd. Xanthochromic fluid [xf]; acute/early-subacute blood level [b]; fourth ventricle [fv]; haemosiderin due to old (>30 d) bleeding [h]; solid component of the tumour [st]; basilar artery [ba].

tion on the outcome or the histopathology of the lesion and of course we cannot exclude a much rarer pre-geniculate schwannoma of the facial nerve that may mimic a more common VS both clinically and at neurophysiological examination. Moreover, all the radiological findings we identified and described are characteristic for a tumour arising from Schwann cells and is therefore unlikely to be a lesion of a different nature. Since current guidelines suggest

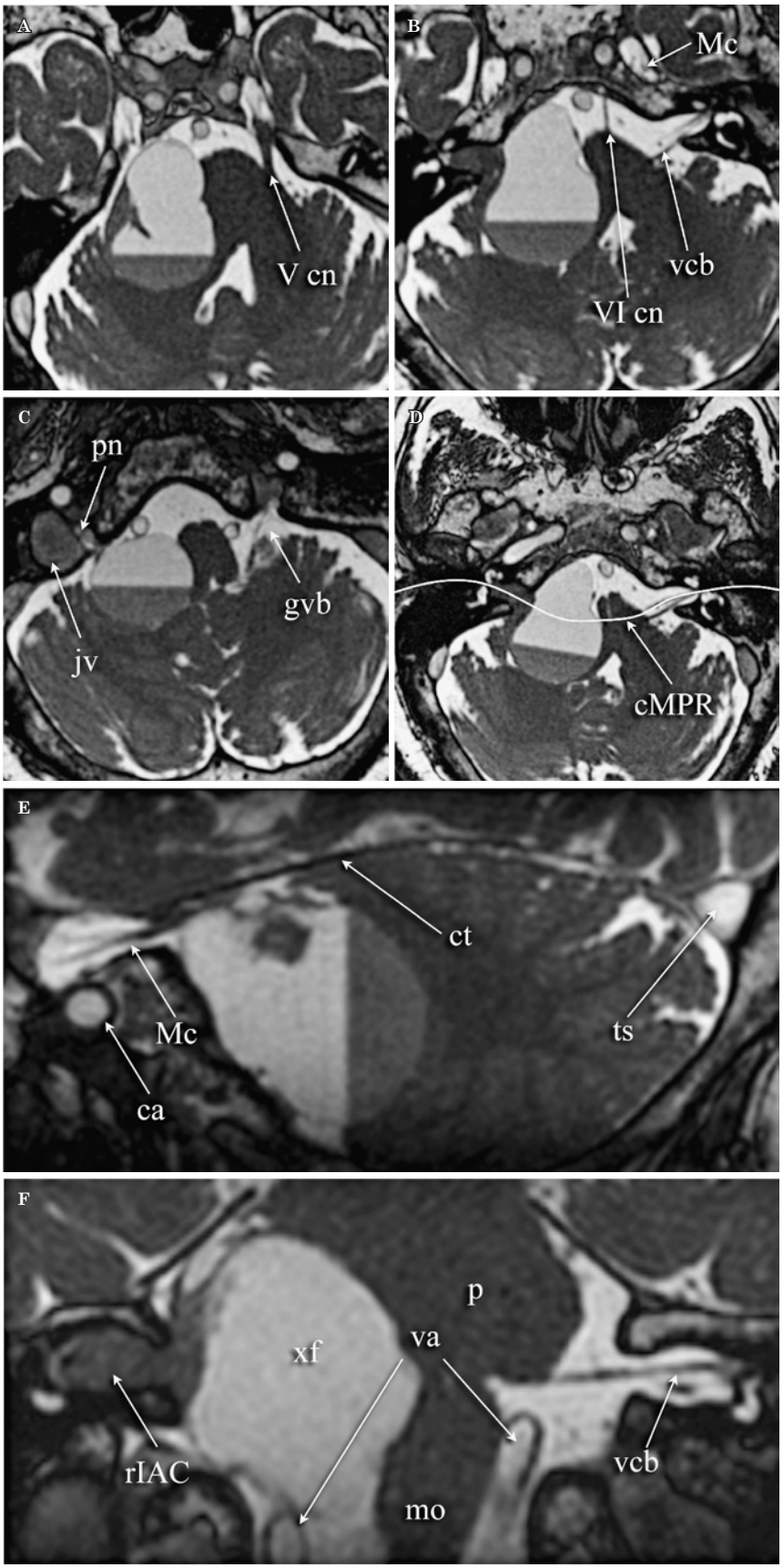

Figure 2 Axial (A-C) and sagittal (E) MPR images from the volumetric FIESTA sequence (0.8 mm isotropic, heavily T2-weighted acquisition). F) Sagittal curved MPR [cMPR] obtained from the plane shown in (D). V cn: left trigeminal nerve leaving the pons [p] and entering Meckel's cave [Mc]. The right preganglionic trigeminal nerve was strongly rostral-medially displaced (trigeminal ganglion and all the major divisions inside the right Mc were normal). VI cn: left abducens nerve entering Dorello's canal with a normal extension (not shown) beneath Grüber's ligament up to and across the cavernous sinus. Normal left glossopharyngeal and vagus nerve branches [gvb]; right branches are displaced since the mass extends caudally up to obstructing the ostium of the pars nervosa [pn] of the jugular foramen; jugular vein [jv] inside the pars vascularis of the foramen. Vestibulo-cochlear branches [vcb]; cerebellar tentorium [ct]; transverse sinus [ts]; carotid artery [ca]; vertebral artery [va]; medulla oblongata [mo].

a "watch and wait" option instead of surgical treatment especially for the geriatric population and "small" VSs, it seems fair to keep in mind the possible unusual clinical presentation due to an acute intratumoral haemorrhage that may require immediate hospitalization.

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